

For Research Use Only

NPC1 Recombinant monoclonal antibody, PBS Only

Catalog Number:86233-5-PBS



Basic Information

Catalog Number: 86233-5-PBS	GenBank Accession Number: BC063302	Purification Method: Protein A purification
Size: 100ug , Concentration: 1 mg/ml by Nanodrop;	GeneID (NCBI): 4864	CloneNo.: 250854D2
Source: Rabbit	UNIPROT ID: O15118	
Isotype: IgG	Full Name: Niemann-Pick disease, type C1	
Immunogen Catalog Number: AG4946	Calculated MW: 142 kDa	
	Observed MW: 160-200 kDa	

Applications

Tested Applications:
WB, IF/ICC, Indirect ELISA

Species Specificity:
human

Background Information

Niemann-Pick Type C (NPC) disease is a lysosomal storage disorder characterized by the accumulation of unesterified cholesterol and other lipids in the endolysosomal system. NPC disease results from a defect in either of two distinct cholesterol-binding proteins: a transmembrane protein, NPC1, and a small soluble protein, NPC2. NPC1 or NPC2 deficiency models showed that the functions of these two proteins within lysosomes are linked closely. NPC1 is a typical transmembrane protein and contains a number of modification sites for glycosylation. Defects in NPC1 are the cause of Niemann-Pick disease type C1 which exhibits highly variable clinical phenotype. Moreover, NPC1 may play a role in vesicular trafficking in glia, a process that may be crucial for maintaining the structural and functional integrity of nerve terminals.

Storage

Storage:
Store at -80°C.

Storage Buffer:
PBS only, pH7.3

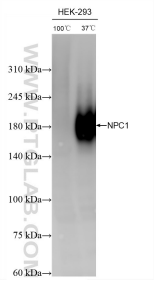
For technical support and original validation data for this product please contact:

T: 1 (888) 4PTGLAB (1-888-478-4522) (toll free in USA), or 1(312) 455-8498 (outside USA)

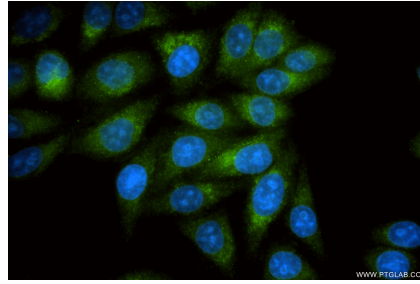
E: proteintech@ptglab.com
W: ptglab.com

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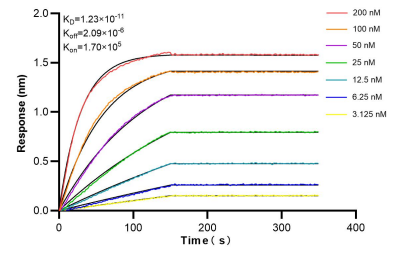
Selected Validation Data



100°C incubated and 37°C incubated HEK-293 cell lysates were subjected to SDS PAGE followed by western blot with 86233-5-RR (NPC1 antibody) at dilution of 1:5000 incubated at room temperature for 1.5 hours. This data was developed using the same antibody clone with 86233-5-PBS in a different storage buffer formulation.



Immunofluorescent analysis of (-20°C Methanol) fixed HepG2 cells using NPC1 antibody (86233-5-RR, Clone: 250854D2) at dilution of 1:400 and CoraLite® 488-Conjugated Goat Anti-Rabbit IgG(H+L) (SA00013-2). This data was developed using the same antibody clone with 86233-5-PBS in a different storage buffer formulation.



Bi-layer interferometry (BLI) kinetic assays of 86233-5-RR against Human NPC1 were performed. The affinity constant is 12.3 pM.